Case presentation

Abdelrahman Amer

Resident of Rheumatology & Rehabilitation

Male patient named Mohamed Ali aged 29 years old from Mansoura works as a doctor. Married with one offspring with no special habits.

Main complaint is Fever, Fatigue, weight loss

The condition started 1 year ago by gradual onset and intermittent course of Flu like symptoms ,
Fever (More than 39 &continuous over day and night) , sore throat , runny nose , night sweating , malaise, generalized bone aches and muscle aches followed by subsequent weight loss of 15kgs over past year
(1st attack lasted 35 days then intermittent course then current attack lasting since 5 months).

Patient took Augmentin /Ciprofloxacin /Tamiflu/Immuguard & fever did not improve.

The patient seeked medical advice and went to tropical medicine consultant who suspected Sarcoidosis
So gave him2 amp of dexamethasone then solupred 10mg/day with good improvement of fever

- No swollen nor red nor hot joints
- Knee and ankle pain
- No ms weakness nor tenderness
- No spine pain

- Oral ulcers (lips and cheeks w are multiple and painful)
- Swollen and painful gums
- Sore throat

- Swelling in neck and axilla
- Swelling submandibular and sunlingual glands in 1st attack and parotid gland in 2nd attack
- Skin rash over back of arm (red , raised and tender)
- No (Alopecia , photosensitivity, Malar rash, Raynaud phenomena, digital ulcers , psoriasis , skin tightness , SC nodules , Nail changes)

- No hearing affection nor nose bleeding
- No dysphagia
- No dryness of mouth
- No hoarseness
- Eye shows no redness nor blurred vision nor pain nor dryness
- No headache, No dizziness , No seizures , No psychosis, No parathesia
- No anxiety, No depression, No sleep disturbance

- No shortness of breath no cough no expectoration no hemoptysis no wheezing

- Nausea , heart burn , No vomiting . No diarrhea nor constipation , No Jaundice

- No Genital ulcers nor genital rash No abnormal discharge from penis
- No dysuria no change in color of urine
- No DM No HTN No thyroid disease

Current medications

- Drug allergy to Metronidazole
- Solupred 40mg/d partially effective
- HCQ 400mg/d
- Imuran 100mg/d
- MTX 12.5mg/week
- Paracetamol 2gm/day
- Diclofenac Sodium 150mg/day

Past history:

- No previous attacks
- No other immunological conditions
- No surgical operations
- No blood transfusion
- No hospital admission
- No past medical hx of (TB , Asthma , pneumonia , jaundice, cancer)

Past medications

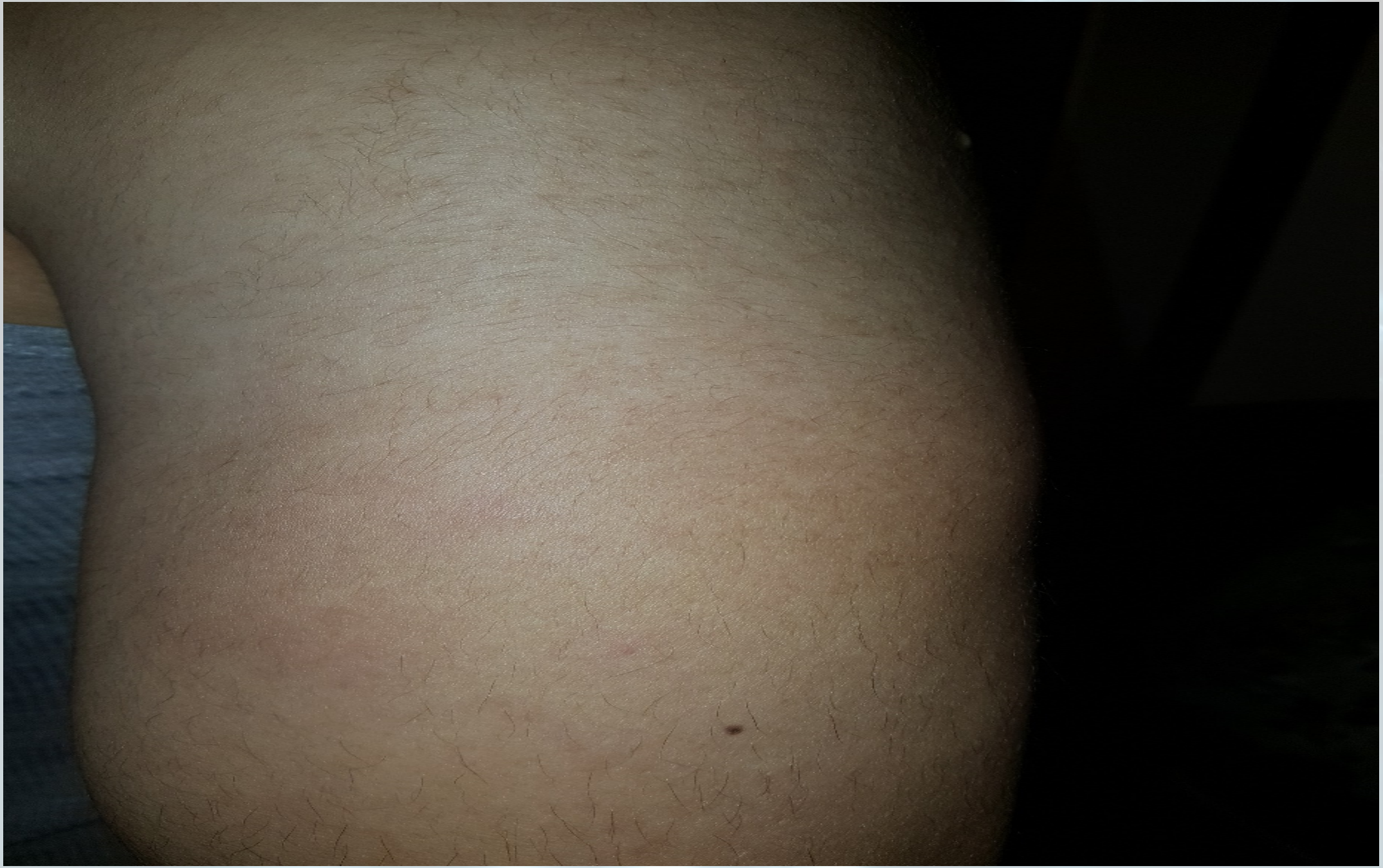
- Dexa amp (many many times in last year)
- Solupred 5-10mg/d at 1st attack w was effective to suppress fever

Family history

- Grand mother RA
- No similar condtion
- No consanguinity

General examination

- Patient appears ill , oriented to time , place and persons
- Vital signs Temp 39.5, pulse 70, BP 120/80
- Height 1.9m Wt 94kg
- Complexion no pallor no jaundice no cyanosis
- Head and neck : oral ulcer single and painful
- Parotid gland enlarged and tender
- LN : single supraclavicular LN enlarged firm tender mobile
- Skin rash over back of arms : reddish , raised and painful
- No abnormal mass
- Normal exam of chest and abdomen



Musculoskeletal examination

- No tender , red , hot ,swollen joints
- Normal range of motion of joints

Spine exam

- Inspection : free
- Palpation : mild tenderness over c7
- Normal range of motion

Neurological exam

Motor :

- Intact ms power , ms tone
- Normal reflexes

Sensory:

- Intact deep and superficial sensation

Laboratory Investigations

CBC	2/2018	1 /2018	3/2017	4/2017
HB	14	13.6	13.7	13
MCV	89	90	90	94
MCH	28	28	28	27
Platelet	276	290	328	542
TLC	2.9 low	1.9 low	6.1	11.5 high
Neutrophils	44.8% low	21.4% low	56%	59%
lymphocytes	41% relative incr	61% relative incr	31%	27%
Monocytes	12% Normal	15% relative incr	12.3% relative incr	13% absolute monocytosis
Eosinophils	1.7%	2.6%	0.3%	1%

Blood film

Comment on blood film:

Mild normocytic normochromic anemia , normal platelets count and morphology , normal WBC count however neutrophil excess toxic granulation suggesting pyogenic infection or inflammatory condition

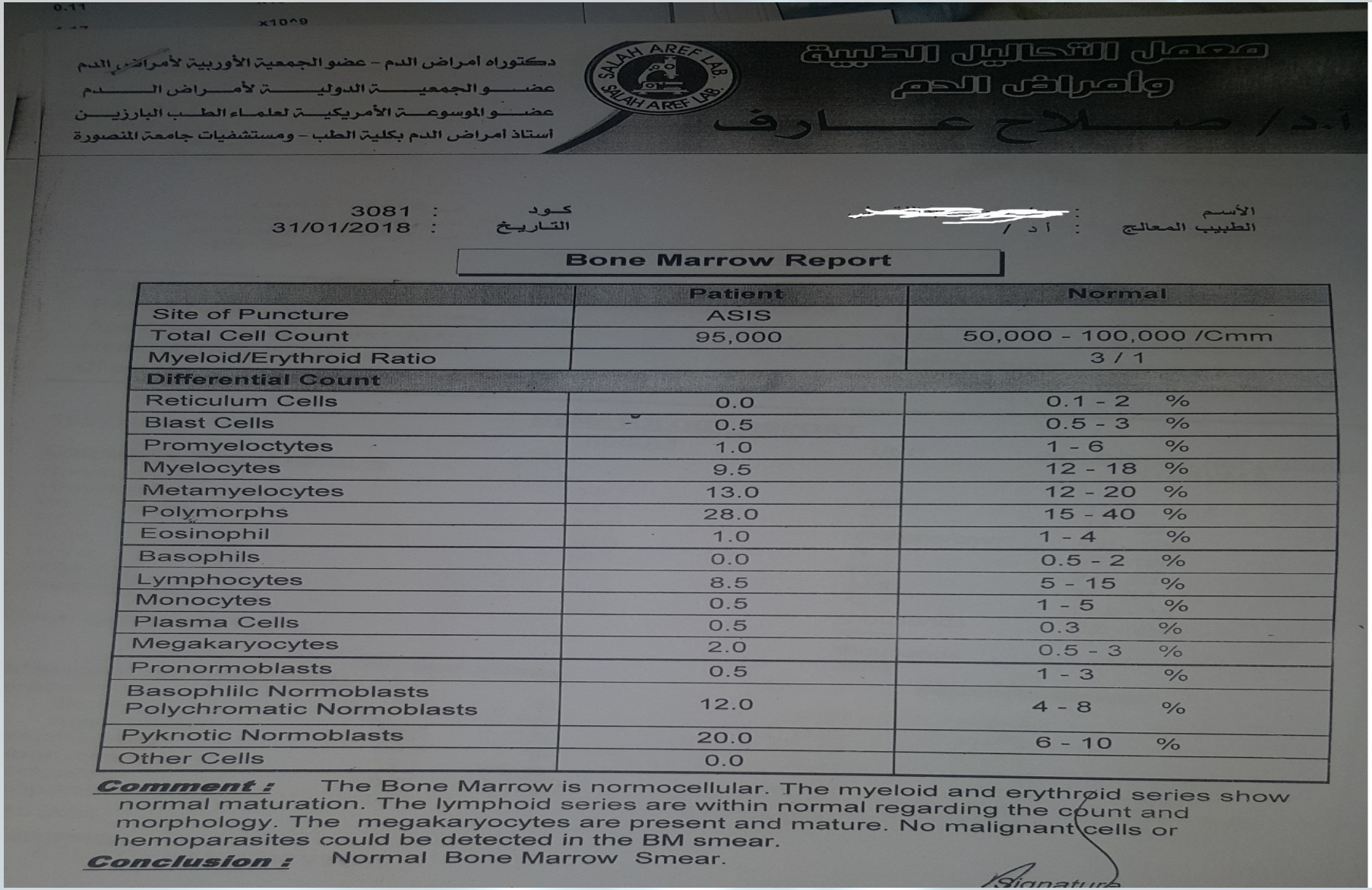
	3/2017	4/2017	1/2018	2/2018
ESR	90/120	67/112	58/90	28/57
CRP	+VE 78	+ve 10	2.5	

ANA	**-VE**
AntidsDNA	-VE
RF	-VE
SERUM TSH	0.6
SGPT	28
S.Creatinine	1

CMV IgM/IgG	-ve
IMN (Monospot test)	-ve
Widal test	-ve
Tuberculin test	-ve
HBsAG	-ve
HCV Ab	-ve
HIV Ab	-ve
blood culture	No growth for aerobic and anaerobic organisms

Serum uric acid	**5.9 mg/dl (Normal)**
Serum TSH	0.63 Uiu/ml (Normal)
Serum ferritin	445.1 ng/ml (NRR15 30-400)
Serum LDH	282 U/L (NRR 120-246)
Urine analysis	No pus cells Normal except few amorphous urate and few calcium oxalate
Lipid profile	High TG 165 Normal total cholesterol 156 Normal LDL 103 Normal HDL 20 High VLDL Cholesterol 33 HDL Risk factor 7.8

Bone marrow aspiration



معمل التحاليل الطبية وأمراض الدم

أ.د/ صلاح عارف

دكتوراه أمراض الدم – عضو الجمعية الأوربية لأمراض الدم
عضو الجمعية الدولية لأمراض الدم
عضو الموسوعة الأمريكية لعلماء الطب البارزين
أستاذ أمراض الدم بكلية الطب – ومستشفيات جامعة المنصورة

SALAH AREF LAB.

الأسم : [redacted]
الطبيب المعالج : أ د /

كود : 3081
التاريخ : 31/01/2018

Bone Marrow Report

	Patient	Normal
Site of Puncture	ASIS	
Total Cell Count	95,000	50,000 - 100,000 /Cmm
Myeloid/Erythroid Ratio		3 / 1
Differential Count		
Reticulum Cells	0.0	0.1 - 2 %
Blast Cells	0.5	0.5 - 3 %
Promyeloctytes	1.0	1 - 6 %
Myelocytes	9.5	12 - 18 %
Metamyelocytes	13.0	12 - 20 %
Polymorphs	28.0	15 - 40 %
Eosinophil	1.0	1 - 4 %
Basophils	0.0	0.5 - 2 %
Lymphocytes	8.5	5 - 15 %
Monocytes	0.5	1 - 5 %
Plasma Cells	0.5	0.3 %
Megakaryocytes	2.0	0.5 - 3 %
Pronormoblasts	0.5	1 - 3 %
Basophlilc Normoblasts Polychromatic Normoblasts	12.0	4 - 8 %
Pyknotic Normoblasts	20.0	6 - 10 %
Other Cells	0.0	

Comment : The Bone Marrow is normocellular. The myeloid and erythroid series show normal maturation. The lymphoid series are within normal regarding the count and morphology. The megakaryocytes are present and mature. No malignant cells or hemoparasites could be detected in the BM smear.

Conclusion : Normal Bone Marrow Smear.

Signature

- The bone marrow is normocellular. The myeloid and erythoid series show normal maturation.
- The lymphoid series are within normal regarding count & morphology.
- The megakaryocytes are present & mature

- No malignant cells or hemoparasites could be detected in bone marrow smear

- Conclusion: normal bone marrow smear

Radiological Investigation

CT chest and abdomen with contrast:

Normal except reactive axillary LNs

Neck US

ULTRASONOGRAPHY OF THE NECK REVEALED :

THE THYROID GLAND

- Normal size . All parts of the gland show normal echo pattern with regular & smooth outline . Normal vascular supply .

THE LARYNX & THE PARALARYNGEAL TISSUE :

- No abnormal mass .
- Normal movements of the vocal cords.

THE SUBMANDIBULAR & THE PAROTID SALIVARY GLANDS:

- Normal on both sides.

THE LYMPH NODES :

- Single enlarged node (1.7 x 0.9 cm.) is seen in the left supraclavicular region. It shows no obvious hilum .
- Two similar nodes {1.8 x 1.0 & 1.0 x 0.9 cm.} are seen inside the left parotid gland.
- Enlarged nodes are seen in both submandibular & both upper deep cervical regions . They show normal shapes & well defined hila.

CONCLUSION:

- Enlarged lymph nodes with no obvious hila are seen in the left parotid & supraclavicular regions... Strict follow up or biopsy is recommended.

Enlarged LNs with no obvious hila are seen in Left parotid & supraclavicular region…strict follow up or biopsy is recommended

LN Biopsy

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Mansoura University
Oncology Center
Oncology Center

جامعة المنصورة
مركز الأورام
مركز الأورام

Patient ID	90000000701458	Patient Name	[redacted]
Patient Sex	Male	Patient Age	

Pathology, 5/4/2017 11:37 AM, Surgical day care ward

Pathology Number	1709/6-4-2017/biopsy
Clinical History	29 years old male patient with ? sarcoidosis underwent excision of left supraclav tender LN about 1.7 cm
Gross Appearance	Fragmented tissue measures 2 x 1 cm in aggregate , firm whitish , totally processed
Microscopic Examination	_ Sections prepared revealed nodal tissue with areas of necrosis associated with infiltration by inflammatory cells , macrophages and abundt karyorhectic debris.
Diagnosis	_ Narcotizing lymphadenitis
Doctor	Wageha Kandil
Doctor	Amr EL-Karef
Doctor	Mai Ali
Doctor	

Summary

Male patient 29ys old presented with

- Fever (persistent over day and night and over 39 Respond partially to steroid , NSAID, Paracetamol and not respond to antibiotics and Tamiflu) , malaise , fatigue , wt loss
- Oral ulcers & pharyngitis & Gingivitis
- Lymphadenopathy (cx and axillary) w is mobile, firm & tender
- Salivary gland swelling (parotid , sublingual & submandibular)
- Skin rash over upper arm (red ,raised & tender)
- Leucopenia and neutropenia
- High ESR & CRP
- Normal (LFT , KFT ,TSH,Urine analysis ,blood culture , Blood film , BMA, CT chest and abdomen)
- -ve(ANA , AntidsDNA , RF , CMV , IMN , HBC, HCV, HIV , Widal T , Tuberculin T)
- Mildly elevated S.ferritin & LDH

Differential diagnosis:

- Lymphoma
- Leukemia
- Castleman disease
- Infections (TB, EBV ,CMV, IMN, lymphogranuloma venereum , Toxoplasma)
- SLE
- Sarcoidosis
- Adult onset still disease
- Kawasaki disease
- PFAPA syndrome (periodic fevers with aphthous stomatitis, pharyngitis, and adenitis)
- Cyclic neutropenia

Not Still ‘s disease

- Fever not typical pattern (quotidian fever)
- Skin rash is non specific (Nononpruritic macular or maculopapular skin rash that is salmon-colored in appearance over the trunk or extremities during febrile episodes)
- CBC : leucopenia not leukocytosis
- S. Ferritin mild increase

Not Leukemia and lymphoma

Excluded by blood film , BMA , LN biopsy

Not SLE

ANA-ve 3 times over course of illness

Not Sarcoidosis

- CT chest normal (No opacities nor hilar lymphadenopathy)
- LN Biopsy shows no noncaseating granulomas

PFAPA syndrome (Marshall syndrome)

Diagnostic criteria — To diagnose PFAPA syndrome, all of the following should be present (at any age):

- More than three documented episodes of fever, lasting no more than five days and occurring at regular intervals (For individual patients, intervals between attacks are nearly identical within a range of three to six weeks, and the symptoms with each episode are identical).
- Pharyngitis plus tender cervical lymphadenopathy or aphthous ulcers.
- Normal growth parameters and good health between episodes.
- Prompt resolution of symptoms with a single dose of prednisone

Exclusion criteria — The presence of any of the following features, precludes the diagnosis of PFAPA syndrome:

- Neutropenia — Neutropenia immediately preceding or during an attack suggests cyclic hematopoiesis, the disease that resembles PFAPA most closely.
- Atypical symptoms — PFAPA attacks have stereotypical symptoms that are easily recognizable by caretakers. The following symptoms as a part of most attacks should trigger consideration of other diagnoses: cough, coryza, severe abdominal pain, significant diarrhea, rash, arthritis, or neuromuscular symptoms. Genetic testing for autoinflammatory diseases should be considered for patients with the above symptoms, except cough and coryza
- Elevated acute phase reactants between attacks — Laboratory evidence of a persistently elevated acute phase protein response (eg, elevated ESR or CRP) suggests a chronic illness with intermittent flares rather than a true periodic disorder.
- Family history of recurrent fever — A family history of periodic fevers should lead to consideration of one of the hereditary autoinflammatory syndromes

Not PFAPA

- Fever not typical pattern
- No Prompt resolution of symptoms with a single dose of prednisone
- CBC: Usually leukocytosis and normal in between attack

Neutropenia is exclusion criteria

Cyclic neutropenia

- Disorder with recurrent fevers that last 5 to 14 days, recurs every 21 to 35 days, and coincides with episodic neutropenia (neutrophils ≤500/mm3).
- Common features include fatigue, pharyngitis, oral ulcers, stomatitis, cellulitis, and lymphadenopathy.
- **Diagnosis** — The diagnosis of cyclic neutropenia is established by documentation of an absolute neutrophil count <200/microL on at least three to five consecutive days per cycle of each of three regularly spaced cycles and a nadir <200/microL. This requires measurement of the ANC at least twice a week for a minimum of six and preferably eight consecutive weeks. The absolute neutrophil count in general should not be above2000/microL at other times

- Such patients are at risk for infection and sepsis and this auto-somal dominant disorder results from a mutation in the neutrophil elastase gene (ELA-2 or ELANE).
- Cyclic neutropenia differs from congenital neutropenia, which has more profound neutropenia and greater risk of infection.
- Effective treatments include granulocyte colony-stimulating factor (GCSF), steroids and, in some cases, cyclosporine

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Member, United States Canadian Academy of Pathology

دكتور
ايليا انيس اسحق
استاذ الباثولوجيا - كلية طب القصر العيني
زميل معهد الباثولوجيا للقوات المسلحة الأمريكية
عضو أكاديمية الباثولوجيا للولايات المتحدة و كندا

Patient Name : [redacted]
Analysis ID : 273267
Age : 29 Y
Gender : Male
Referred By Prof. Dr. : [redacted]
Clinical Diagnosis : Histiolytic necrotizing lymphadenitis
Nature of Specimen : Referred block
Receiving Date : 25-02-2018
Delivery Date : 27-02-2018

IMMUNOHISTOCHEMISTRY REPORT

Technique :

Sections were prepared from the paraffin block then treated by monoclonal antibodies and detection kit (EnVision FLEX) using DAB as chromogen and hematoxylin as counter-stain, using Omnis machine.
The sections were treated against
- CD 20
- CD 3

Results :

CD 20 showed positive small aggregates widely spaced by necrosis.
CD 3 showed many positive small lymphocyte.

Conclusion :

Referred block, FINDINGS ARE COMPATIBLE WITH REACTIVE NODAL TISSUE WITH WIDELY SPACED NECROSIS, COMPATIBLE WITH KIKUCHI DISEASE.

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Kikuchi disease

or

Kikuchi-Fujimoto disease

or

Kikuchi histiocytic necrotizing lymphadenitis

pathogenesis

- unknown, the clinical presentation, course, and histologic changes suggest an immune response of T cells and histiocytes to an infectious agent proposed, (EBV) human herpesvirus 6 (HIV), parvovirus B19 , paramyxoviruses, parainfluenza virus, *Yersinia enterocolitica,* and *Toxop/asma.*

- A possible role for interferon-gamma and interleukin (IL)-6 in the pathogenesis of this syndrome is suggested by one study of four men with biopsy-proven Kikuchi disease [ll]. During the acute phase of illness, these patients had elevated serum levels of interferon-gamma andIL-6 but not interferon-alpha, tumor necrosis factor, orIL-2. The interferon-gamma and IL-6 levels returned to normal during convalescence. Subsequent reports support this role [12.]. Analysis of lymph node biopsies in patients with Kikuchi disease show T-bet-expressing CD4 cells accompanied by T-bet positive CDS and B cells [13].

EPIDEMIOLOGY

- While initially described in young women, Kikuchi disease clearly also occurs in men.

- The ratio of affected males to females was 1:4

- Most patients are younger than 40 years of age,reported in patients ranging in age from 6 to 80 years, most of whom were previously well . The mean age at presentation in a United States series was 30 years .

Ethnic distribution

Although initially described in two separate cases from Japan, Kikuchi disease has since been found in all racial and ethnic groups and in many countries, including the United States

CLINICAL FEATURES

- The most common clinical presentation of Kikuchi disease is fever and cervical lymphadenopathy in a previously well young woman.
- Fever is a primary symptom in 30-50%. It is typically low grade and persists for about one week , rarely for up to one month.
- In one study of 86 children with Kikuchi disease, the median duration of fever was 9 days but was more prolonged in those with higher fever (39.0°C), leukopenia, and larger lymph nodes

Lymphadenopathy	100%
Fever	35%
Rash	10%
Arthritis	7%
Fatigue	7%
Hepatosplenomegaly	3%

- Rigors, myalgia, arthralgia, and chest and abdominal pain

- Night sweats, nausea, vomiting, diarrhea, and weight loss have also been reported and appear to be more prominent in patients with extra nodal disease

- There have been several reports of disease limited to the mediastinum, with no cervical involvement. Mesenteric lymphadenitis may mimic appendicitis

Rash

- Transient skin rashes similar to rubella or drug-induced eruptions may be seen in sicker patients may be pruritic
- Some reports describe skin manifestations in up to 40 % including facial erythema; erythematous macules, patches, papules, orplaques; scattered indurated lesions; ulcers; polymorphous light eruptions; leukocytoclastic vasculitis; scales; pruritus; alopecia; conjunctival injection; and oral ulceration

- The presence of a malar "butterfly rash" should raise the diagnosis of SLE that, as noted above, has been associated with Kikuchi disease.

Lymph node involvement

- Lymph node involvement is usually cervical and localized in Kikuchi disease

- may be more extensive node involvement or involvement of other sites. These sites include the axillary, epitrochlear, mediastinal, mesenteric, inguinal, intraparotid, iliac, retrocrural, celiac, and peripancreatic nodes.
- The nodes are usually only moderately enlarged (1 to 2 cm in diameter) but occasionally are much larger (5- 7 cm) .
- They are typically firm, smooth, discrete, and mobile.
- The nodal enlargement is often associated with dull or acute pain.

Other manifestations

- Aseptic meningitis , meningoencephalitis , acute cerebellar symptoms with tremor and ataxia
- thyroiditis and parotid enlargement ,
- pleural effusions
- polymyositis, hemophagocytosis
- panuveitis , bilateral papillary conjunctivitis
- autoimmune hepatitis brachia! neuritis , and peripheral neuropathy
- There are reports of antiphospholipid syndrome with multiorgan failure

LABORATORY STUDIES

- The majority have a normal CBC , although leukopenia is seen in up to 43 %
- Atypical lymphocytes have been reported in up to 25 %.
- Other reported findings include thrombocytopenia, pancytopenia, and in those with severe disease, anemia of chronic disease

- ESR can be normal but was elevated to more than 60 mm/hour in 70% of patients in one series

- Other nonspecific findings can include mildly abnormal liver function tests and elevated serum lactate dehydrogenase.

Bone marrow examination

- An increase in macrophages without atypical cells is the most frequent bone marrow

Serologic studies

- ANA, rheumatoid factor, and lupus erythematosus preparations are generally negative.
- Some patients initially diagnosed with Kikuchi disease have subsequently developed systemic lupus erythematosus (SLE) , and an ANA test should be performed in patients with suspected Kikuchi syndrome who have features suggestive of SLE in order to exclude this diagnosis
- Serology for Epstein-Barr virus, cytomegalovirus, human immunodeficiency virus (HIV), toxoplasmosis, *Y. enterocolitica* , cat scratch disease, and other infectious agents is often performed since these infections are considered in the differential diagnosis of fever and lymphadenopathy.

DIAGNOSIS

The diagnosis of Kikuchi disease is made by lymph node biopsy. Biopsy should be performed, despite the self-limited nature of this syndrome, in order to exclude more serious conditions requiring aggressive therapy such as lymphoma. Patients with Kikuchi disease have been misdiagnosed as having lymphoma and treated with cytotoxic agents when physicians and pathologists are unfamiliar with this entity

IMAGING STUDIES

- Computed tomography (CT) imaging of the affected lymph nodes typically shows perinodal infiltration (Sl percent) and homogenous nodal contrast enhancement (S3 percent) [Sl]. Nodal cortical attenuation and its ratio to adjacent muscle on CT imaging may be used to differentiate Kikuchi disease from nodal reactive hyperplasia and tuberculous lymphadenopathy, if the lymphadenopathy is not necrotic. If the lymphadenopathy is necrotic, the pattern of necrosis may allow differentiation from tuberculosis .

- On ultrasound, lymph nodes may appear suspicious for malignancy

TREATMENT

- No effective treatment has been established for Kikuchi disease.
- Signs and symptoms usually resolve within one to four months
- Patients with severe or persisting symptoms have been treated with glucocorticoids
- Affected patients should be followed for some years because they can develop systemic lupus erythematosus, and recurrences of Kikuchi disease can occasionally continue for many years

Kikuchi-Fujimoto's disease associated with systemic lupus erythematous: difficult case report and literature review.

Review article

Ruaro B, et al. Lupus. 2014.
Show full citation

Abstract

Kikuchi-Fujimoto's disease (KFD), or histiocytic necrotizing lymphadenitis, is a benign and self-limiting disease of unknown aetiology. KFD tends to affect a young population under 30 years of age and predominantly females. KFD is a rare pathology and its association with systemic lupus erythematosus (SLE) is not frequent. Herein, we present the case of a male Italian patient with SLE in association with KFD with 5 years of follow-up, where a differential diagnosis from infection or lymphoproliferative disease was problematic.

PMID: 24739458 [PubMed - indexed for MEDLINE]

[Kikuchi-Fujimoto's disease and adult-onset Still's disease. A rare co-occurence].

Sondermann W, et al. Hautarzt. 2015.
Show full citation

Abstract

Kikuchi-Fujimoto's disease and adult-onset Still's disease are rare inflammatory conditions with overlapping clinical features. Adult-onset Still's disease causes high fevers, a typical salmon-colored rash, and joint pain. The principal symptom of Kikuchi's disease is cervical lymphadenopathy with typical histopathological features including extensive necrosis of the involved lymph nodes. Here, we report on a rare case of concurrent adult-onset Still's disease and Kikuchi-Fujimoto syndrome in a young Caucasian patient.

PMID: 26115972 [PubMed - indexed for MEDLINE]

Full text

Infect Chemother. 2016 Jun; 48(2): 127–131.
Published online 2016 Jun 30. doi: 10.3947/ic.2016.48.2.127
PMCID: PMC4945722

Recurrent Kikuchi's Disease Treated by Hydroxychloroquine

Miri Hyun,[1] In Tae So,[1] Hyun Ah Kim,[1] Hyera Jung,[2] and Seong-Yeol Ryu[1]

Author information ▶ Article notes ▶ Copyright and License information ▶

Abstract

Go to:

Kikuchi-Fujimoto disease (KFD) is a benign, self-limiting disease, with a specific histopathology. It can be diagnosed clinically, and specific symptoms include fever and cervical lymphadenopathy. The histological finding of KFD in cervical lymph nodes includes necrotizing lymphadenitis. KFD needs conservative treatments. If KFD persists for a long period, steroids or nonsteroidal antiinflammatory drugs can be used to control symptoms. Previous studies have reported the treatment of KFD with hydroxychloroquine (HC) in patients unresponsive to steroids. Herein, we report a case of a 25-year-old female patient diagnosed with KFD unresponsive to steroids, and was successfully treated with HC.

Singapore Med J. 2010 Jan;51(1):e18-21.

Severe Kikuchi's disease responsive to immune modulation.

Lin DY[1], Villegas MS, Tan PL, Wang S, Shek LP.

Author information

Abstract

Kikuchi's disease, although an uncommon entity, has been increasingly reported since it was first discovered in 1972. The most common manifestation of Kikuchi's disease, cervical lymphadenopathy, has no clinically distinguishable features. Therefore, a diagnosis of Kikuchi's disease has largely been based on clinical suspicion and histopathological confirmation. We present a 15-year-old Chinese girl with severe Kikuchi's disease, whose relapsing course was only responsive to highdose steroid and intravenous immunoglobulin therapy.

- IV methyl prednisolone300 mg (7.5 mg/kg) was administered
daily over three days, followed by a dose of IV IgG 15 mg (0.4 mg/kg).

- This was repeated one month later with continued good response.

- The patient demonstrated an almost complete clinical response to the first two doses, and there was no requirement to administer another course.